# General

## Guideline Title

Ependymomas.

## Bibliographic Source(s)

Alberta Provincial CNS Tumour Team. Ependymomas. Edmonton (Alberta): CancerControl Alberta; 2012 May. 8 p. (Clinical practice guideline; no. CNS-004). [27 references]

## Guideline Status

This is the current release of the guideline.

# Recommendations

## Major Recommendations

World Health Organization (WHO) Grade II Ependymomas

1. Surgery represents the standard initial treatment. Maximal surgical resection is important whenever possible.
2. Postoperative magnetic resonance imaging (MRI) within 72 hours allows assessment of residual tumour and guides further management.
3. Postoperative radiotherapy may be considered for a known or suspected residual intracranial tumour, in order to increase local disease control. If administered, a standard dose of 45–54 Gy may be administered in 1.8–2.0 Gy per fraction.
4. Currently, there is no evidence that the addition of chemotherapy to surgery or radiotherapy improves outcome.

WHO Grade III (Anaplastic) Ependymomas

5. Surgery plus radiotherapy represents the standard treatment.
6. Postoperative radiotherapy doses of 54–60 Gy should be administered in 1.8–2.0 Gy per fraction whenever possible. The dose to the optic chiasm, optic nerves, and spinal cord should be limited as appropriate.
7. Craniospinal irradiation in patients with evidence of craniospinal spread should be considered. A palliative approach, using limited doses and volumes of radiotherapy or other approaches may be used.
8. Chemotherapy is a treatment option being evaluated; recurrent patients should be considered as candidates for chemotherapy or clinical trials.

Follow-up

9. Close observation and long-term follow-up is recommended for all patients with ependymomas, due to late effects of radiotherapy in long-term survivors.

## Clinical Algorithm(s)

None provided

# Scope

## Disease/Condition(s)

Ependymomas (intracranial and spinal cord)

## Guideline Category

Management

Treatment

## Clinical Specialty

Neurological Surgery

Neurology

Oncology

Radiation Oncology

## Intended Users

Advanced Practice Nurses

Nurses

Physician Assistants

Physicians

## Guideline Objective(s)

To provide optimal treatment strategies for adult patients with World Health Organization (WHO) grades II and III (anaplastic) ependymomas

## Target Population

Adults over the age of 18 years with World Health Organization (WHO) grades II and III (anaplastic) ependymomas

Note: Different principles may apply to pediatric patients.

## Interventions and Practices Considered

1. Surgical resection (gross total resection)
2. Postoperative magnetic resonance imaging (MRI)
3. Postoperative radiotherapy
4. Craniospinal irradiation

5. Chemotherapy
6. Close observation and long-term follow-up

### Major Outcomes Considered

- Survival rates (10-year, 5-year, overall, progression-free)
- Recurrence rate
- Surgical morbidity

## Methodology

### Methods Used to Collect/Select the Evidence

Searches of Electronic Databases

### Description of Methods Used to Collect/Select the Evidence

Research Questions

Specific research questions to be addressed by the guideline document were formulated by the guideline lead(s) and Knowledge Management (KM) Specialist using the PICO question format (patient or population, intervention, comparisons, outcomes).

Guideline Question

- What are the optimal treatment strategies for adult patients with World Health Organization (WHO) grades II and III (anaplastic) ependymomas?

Search Strategy

For the development of the original guideline, medical journal articles were searched using the Medline (1950 to August Week 3, 2009), EMBASE (1980 to August Week 3, 2009), Cochrane Database of Systematic Reviews (3rd Quarter, 2009), and PubMed electronic databases; the references and bibliographies of articles identified through these searches were scanned for additional sources. The search terms included: Ependymoma [MeSH heading], low-grade ependymoma, subependymoma, myxopapillary ependymoma, practice guidelines, systematic reviews, meta-analyses, randomized controlled trials, and clinical trials. Articles were excluded from the review if they: had a non-English abstract, were not available through the library system, were case studies involving less than 5 patients, or were published prior to the year 2000.

For the 2012 update of this guideline, Medline, EMBASE, Cochrane and PubMed were searched using the terms Ependymoma [MeSH heading], low-grade ependymoma, subependymoma, and myxopapillary ependymoma, limited to clinical trials, clinical trials phase I–IV, controlled clinical trials, randomized controlled trials, published from September 2009 to the present (May 25, 2012). Articles were excluded if they had a non-English abstract, were case studies involving less than 5 patients, did not include survival outcomes or were over 50% patients under 18 years old with a median age of less than 18 years. A review of the relevant existing practice guidelines for ependymomas was also conducted by accessing the practice guidelines on the websites of the British Columbia Cancer Agency (BCCA), Cancer Care Ontario (CCO), the National Comprehensive Cancer Network (NCCN), American Society of Clinical Oncology (ASCO), European Society of Medical Oncology (ESMO) and the National Cancer Institute (NCI).

### Number of Source Documents

Not stated

### Methods Used to Assess the Quality and Strength of the Evidence

Not stated

## Rating Scheme for the Strength of the Evidence

Not applicable

## Methods Used to Analyze the Evidence

Systematic Review with Evidence Tables

## Description of the Methods Used to Analyze the Evidence

Evidence was selected and reviewed by a working group comprised of members from the Alberta Provincial CNS Tumour Team and a Knowledge Management (KM) Specialist from the Guideline Utilization Resource Unit (GURU). A detailed description of the methodology followed during the guideline development process can be found in the Guideline Utilization Resource Unit Handbook (see the "Availability of Companion Documents" field).

Evidence Tables

Evidence tables containing the first author, year of publication, patient group/stage of disease, methodology, and main outcomes of interest are assembled using the studies identified in the literature search. Existing guidelines on the topic are assessed by the KM Specialist using portions of the Appraisal of Guidelines Research and Evaluation (AGREE) II instrument (http://www.agreetrust.org) and those meeting the minimum requirements are included in the evidence document. Due to limited resources, GURU does not regularly employ the use of multiple reviewers to rank the level of evidence; rather, the methodology portion of the evidence table contains the pertinent information required for the reader to judge for himself the quality of the studies.

## Methods Used to Formulate the Recommendations

Expert Consensus

## Description of Methods Used to Formulate the Recommendations

Formulating Recommendations

The working group members formulated the guideline recommendations based on the evidence synthesized by the Knowledge Management (KM) Specialist during the planning process, blended with expert clinical interpretation of the evidence. As detailed in the Guideline Utilization Resource Unit Handbook (see the "Availability of Companion Documents" field), the working group members may decide to adopt the recommendations of another institution without any revisions, adapt the recommendations of another institution or institutions to better reflect local practices, or develop their own set of recommendations by adapting some, but not all, recommendations from different guidelines.

The degree to which a recommendation is based on expert opinion of the working group and/or the Provincial Tumour Team members is explicitly stated in the guideline recommendations. Similar to the American Society of Clinical Oncology (ASCO) methodology for formulating guideline recommendations, the Guideline Utilization Resource Unit (GURU) does not use formal rating schemes for describing the strength of the recommendations, but rather describes, in conventional and explicit language, the type and quality of the research and existing guidelines that were taken into consideration when formulating the recommendations.

## Rating Scheme for the Strength of the Recommendations

Not applicable

## Cost Analysis

A formal cost analysis was not performed and published cost analyses were not reviewed.

### Method of Guideline Validation

Internal Peer Review

### Description of Method of Guideline Validation

This guideline was reviewed and endorsed by the Alberta Provincial CNS Tumour Team.

When the draft guideline document has been completed, revised, and reviewed by the Knowledge Management Specialist and the working group members, it is sent to all members of the Provincial Tumour Team for review and comment. This step ensures that those intended to use the guideline have the opportunity to review the document and identify potential difficulties for implementation before the guideline is finalized. Depending on the size of the document, and the number of people it is sent to for review, a deadline of one to two weeks will usually be given to submit any feedback. Ideally, this review will occur prior to the annual Provincial Tumour Team meeting, and a discussion of the proposed edits will take place at the meeting. The working group members will then make final revisions to the document based on the received feedback, as appropriate. Once the guideline is finalized, it will be officially endorsed by the Provincial Tumour Team Lead and the Executive Director of Provincial Tumour Programs.

## Evidence Supporting the Recommendations

### Type of Evidence Supporting the Recommendations

The type of evidence supporting the recommendations is not specifically stated.

## Benefits/Harms of Implementing the Guideline Recommendations

### Potential Benefits

Appropriate management and treatment of ependymomas

### Potential Harms

Gross total resection (GTR) can be hindered by anatomical factors such as adherence of the tumour to surrounding structures, particularly for tumours in the fourth ventricle, brain stem, lower cranial nerves, or major vascular structures. Thus, consideration must be given to the balance between improved survival with GTR and postoperative morbidity related to high-risk surgery.

## Qualifying Statements

### Qualifying Statements

The recommendations contained in this guideline are a consensus of the Alberta Provincial CNS Tumour Team and are a synthesis of currently accepted approaches to management, derived from a review of relevant scientific literature. Clinicians applying these guidelines should, in consultation with the patient, use independent medical judgment in the context of individual clinical circumstances to direct care.

## Implementation of the Guideline

### Description of Implementation Strategy

- Present the guideline at the local and provincial tumour team meetings and weekly rounds.
- Post the guideline on the Alberta Health Services Web site.
- Send an electronic notification of the new guideline to all members of CancerControl Alberta.

# Institute of Medicine (IOM) National Healthcare Quality Report Categories

## IOM Care Need

Getting Better

Living with Illness

## IOM Domain

Effectiveness

# Identifying Information and Availability

## Bibliographic Source(s)

Alberta Provincial CNS Tumour Team. Ependymomas. Edmonton (Alberta): CancerControl Alberta; 2012 May. 8 p. (Clinical practice guideline; no. CNS-004). [27 references]

## Adaptation

Not applicable: The guideline was not adapted from another source.

## Date Released

2012 May

## Guideline Developer(s)

CancerControl Alberta - State/Local Government Agency [Non-U.S.]

## Source(s) of Funding

CancerControl Alberta

## Guideline Committee

Alberta Provincial CNS Tumour Team

## Composition of Group That Authored the Guideline

Members of the Alberta Provincial CNS Tumour Team include medical oncologists, radiation oncologists, neurosurgeons, nurses, neuropathologists, and pharmacists.

## Financial Disclosures/Conflicts of Interest

Participation of members of the Alberta Provincial CNS Tumour Team in the development of this guideline has been voluntary and the authors have not been remunerated for their contributions. There was no direct industry involvement in the development or dissemination of this guideline. CancerControl Alberta recognizes that although industry support of research, education and other areas is necessary in order to advance patient care, such support may lead to potential conflicts of interest. Some members of the Alberta Provincial CNS Tumour Team are involved in research funded by industry or have other such potential conflicts of interest. However the developers of this guideline are satisfied it was developed in an unbiased manner.

## Guideline Status

This is the current release of the guideline.

## Guideline Availability

Electronic copies: Available from the Alberta Health Services Web site .

## Availability of Companion Documents

The following is available:

- Guideline utilization resource unit handbook. Edmonton (Alberta): CancerControl Alberta; 2013 Jan. 5 p. Electronic copies: Available from the Alberta Health Services Web site .

## Patient Resources

None available

## NGC Status

This NGC summary was completed by ECRI Institute on August 12, 2014. The information was verified by the guideline developer on September 22, 2014.

## Copyright Statement



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